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SHORT REPORT

Paget-von Schrötter Syndrome Following Weight Lifting (Effort Thrombosis of Upper Extremity)

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Introduction

Sir James Paget was the first to describe spontaneous thrombosis of upper extremity veins in 1875.[1] Von Schrötter, in 1884, was the first to relate this to thrombotic occlusion of subclavian and axillary veins.[1] This syndrome is relatively rare and accounts for 0.5–1.5% of all venous thrombosis. It is postulated that sheer force could have initiated injury and promoted thrombus formation and local hypercoagulopathy.[2] In some instances, conditions such as a thoracic outlet obstruction predispose to thrombotic events.

Case Report

A 17-year-old Caucasian male with 5 days history of pain, swelling and discoloration of right arm presented to our emergency department. He practiced weight lifting regularly and noted increased swelling after each weight lifting session. In exam, arm and forearm were tender and slightly swollen, but pulse was palpable and strong. Collateral venous distension in chest was noticeable.

Duplex ultrasonography demonstrated thrombosis of subclavian and proximal axillary veins. Digital subtraction venography was performed and revealed complete occlusion of subclavian and axillary vein. Collateral filling was noted in the upper chest and neck with filling of the right jugular vein and right inominate vein.

Mechanical thrombectomy was done with a 6 French Angiojet catheter, resulting in considerable debulking of thrombus within the subclavian vein. Percutaneous balloon angioplasty of the subclavian vein was performed at the first rib-clavicle junction, because of a pattern of high-grade stenosis in the area. Thrombolytic therapy with tissue plasminogen activator (t-pa) was started through a subclavian catheter at 2 mg/h and heparin was started from another site at the rate of 600 units/h. Heparin was changed after 24 h to low molecular weight heparin (1 mg/kg q12 h) which continued for 5 days. About the same time, warfarin was started and was continued to keep the International Normalised Ratio between 2 and 3. Haematological investigation to investigate any possible hypercoagulable state was performed and revealed no abnormality.

The patient was followed up in another centre closer to his home. Six months after the initial event, while he was still on warfarin, he died from an embolic stroke. He refused to give up weight lifting during this time period.

Discussion

Paget-von Schrötter Syndrome usually occurs in young healthy individuals. Symptoms include sudden onset of pain, swelling, non-pitting oedema and cyanosis of the upper extremities.[2] Collateral veins usually are visible around the shoulder and chest wall.

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Pain is exacerbated with exercise, and is improved with rest and limb elevation. This syndrome often follows repetitive activity, such as throwing a ball, house painting, paper hanging, swimming or rowing, and has been called 'effort thrombosis'.[3] New criteria for diagnosing Paget-von Schrötter syndrome include a history of increased upper extremity use before onset of symptoms, the presence of a venographically demonstrated thrombus, and absence of any definable cause.[4]

Zell *et al*.[3] reported 82 patients with subclavian thrombosis, of which 51 had secondary reasons, such as central catheter, carcinoma, postoperative or hypercoagulability, and 31 did not have secondary reasons. Twenty-eight of the 31 patients without secondary reasons showed unusual private or occupational physical exercise as the underlying cause. Sixteen patients reported only short physical exercise and 12 had histories of long hours of exercise.

Venography is the gold standard for diagnosis. Colour duplex has virtually replaced contrast venography in the diagnosis of lower extremity thrombosis, but the opposite is true in the upper extremity.[2] The thoracic outlet abnormalities are usually bilateral, thus both sides should be evaluated. If degree of oedema makes the procedure difficult, digital subtraction techniques can be employed.

Treatment options vary. In short, four treatment modalities should be used: thrombolysis (catheter directed), anticoagulation, angioplasty and stenting (in case of stenosis), and thoracic outlet decompression (early vs. late).

Although Paget-von Schrötter has been associated with sporting activities, its relation with weight lifting has not been mentioned previously and since weight lifting is popular in gyms, close attention and observation of possible correlation can be affecting the health of young otherwise healthy patient population. It seems logical to advise against repetitive upper extremity movements and weight lifting in a patient with Paget-von Schrötter or thoracic outlet syndrome.

References

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